Post-polio Syndrome: An Overview

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Poliomyelitis was a dreaded disease of developed countries, affecting tens of thousands of children and adults during each of the epidemic years up to the mid-1950s. The polio virus is a small RNA virus whose only natural host appears to be man. The vast majority of exposed persons develop either an inapparent infection or a non-specific flu-like illness (non-paralytic poliomyelitis). Secondary invasion of brain and spinal cord is associated with infection and death of motor neurons, with loss of innervation to muscle fibers, and consequent muscle weakness and atrophy. Postmortem studies show that muscle weakness in poliomyelitis is clinically apparent only when more than half of corresponding motor neurons are destroyed. Frequently, muscles can be reinnervated by healthy neighboring motor neurons by a process of axonal sprouting. Thus, partial or complete recovery of muscle bulk and strength can occur, in which subnormal numbers of motor neurons support increased (up to 8-fold) numbers of muscle fibers. It is estimated that about 250,000 people in the United States have survived paralytic poliomyelitis and are alive today.

Recently, it has become clear that some patients who had paralytic poliomyelitis may develop new complaints after decades of stable function. These new symptoms have been designated the “post-polio syndrome” (PPS) or “late sequella of poliomyelitis.” Although some reports of new weakness following polio can be found in the medical literature since 1875, recent epidemiologic studies indicate that new symptoms are common, occurring in approximately 25 percent of patients with antecedent paralytic poliomyelitis. If this estimate is correct, over 50,000 persons in the U.S. are at risk of developing PPS. From published reports, the mean latency of onset has been calculated to be 36 years. Thus, an increasing incidence of new cases will probably continue into the early 1990s, reflecting the last epidemics of the mid-1950s.

The risk of developing PPS appears to correlate with severity of the original poliomyelitis. Thus, a patient with four-limb involvement and a history of respiratory dependence during his polio is more likely to develop new symptoms than a patient with one-limb involvement. The severity of the original onset of polio also seems to predict the latency of developing the syndrome; severely affected patients may develop new symptoms after only 10–20 years, whereas mildly affected patients are more likely to exhibit extended delays in time of onset of PPS.

What causes PPS? Why should a patient who has had stable function for decades develop new symptoms? At the present time, there is little definitive data on this subject. Early conjecture focused on a possible reactivation of the polio virus which had remained latent in the nervous system since the original infection. However, there appears to be little or no evidence for inflammation in post-polio patients; spinal fluid is without the cells, protein, and immunoglobulin which characterize other nervous system viral infections. Some investigators have suggested that the normal attrition of neurons with aging may trigger the post-polio syndrome when superimposed on previous static damage of polio. However, aging-related loss of neurons in the spinal cord normally begins about age 60; the onset of PPS most commonly occurs 30 years after polio and does not correlate with chronological age of the...
Weichers and Hubbel\textsuperscript{8} and Dalakis, et al.\textsuperscript{9} have suggested that motor units grossly enlarged by reinnervation in recovery from poliomyelitis may begin to experience peripheral disintegration with the passage of time. Our own data support this hypothesis in part; late denervation is most common in muscles with the greatest degree of reinnervation. However, we find that group atrophy, a putative indicator of motor neuronal disease (and not terminal axonal degeneration), is also common in patients with prior poliomyelitis.\textsuperscript{10}

Although a bewildering variety of new symptoms are recognized as occurring in PPS,\textsuperscript{4} most new complaints appear to be subsumed under the three major problems of new pain, new weakness, and fatigue (Table 1). Some investigators have theorized that new muscle atrophy and weakness constitutes a separate syndrome, "postpoliomyelitis progressive muscular atrophy" or PPMA.\textsuperscript{9} In this scheme, other symptoms of PPS, such as pain and fatigue, are thought to be manifestations of a separate "musculoskeletal" syndrome due to chronic strain of muscles and joints that have been forced to bear weight in an unnatural fashion.\textsuperscript{11} Common orthopedic deformities in patients with poliomyelitis include knee valgus, varus, and recurvatum, as well as ankle equinus.\textsuperscript{13} However, new weakness can result in new joint instability, and new joint problems may interfere with efficiency of movement. Although a symptomatic approach to separate complaints of PPS patients is warranted, there is little scientific data which supports a division of sub-syndromes of PPS at present. We have found that even patients without new symptoms have evidence of an ongoing neuromuscular disorder.\textsuperscript{10}

New pain is the most common symptom in PPS based on our experience (Table 1), and is a frequent complaint in other series as well.\textsuperscript{4,6} We have evaluated patients experiencing pain in conjunction with an orthoped experienced in neuromuscular disease. Several causes of pain are commonly identified in PPS patients. Perhaps the most common cause is insertionl tendinitis and/or bursitis from chronic overuse and strain of muscle groups with subnormal strength. Palpation of tendons and bursae at common sites of involvement, such as the pes tendon at the medial knee and the trochanteric bursa, will often reveal profound point tenderness consistent with this syndrome. A trial of rest and non-steroidal anti-inflammatory agents may induce remission in this remitting/relapping syndrome. For certain local sites, a steroid injection may be useful; weight reduction and readjustment of weight-bearing (through retraining and/or orthotic devices) may also produce long-range benefits. Degenerative arthritis, found most often in weight-bearing joints (where walking aids are used, the joints of the upper extremities may indeed become weight bearing), may also respond to the same regimen. Nerve compression syndromes characterized by pain and paraesthesias, secondary to positional or repetitive stress, should also be considered in the differential diagnosis of pain in PPS patients.

Another type of pain, unrelated to joint "wear and tear" is muscle pain. This occurs frequently during or after exercise, and may be associated with cramps, fasciculations, or intense local fatigability. This may be related to muscle substitution and/or overwork in denervated muscle, and may ultimately be associated with permanently increased weakness.\textsuperscript{14} Treatment of this muscle pain includes avoiding the circumstances which induce it. Rest, orthoses, or even intermittent wheelchair use should also be considered to reduce load on overworked muscle. Medications which reduce muscle cramps (quinine, diphenylhydantoin) may increase weakness, and should be avoided.

Fatigue is also a common complaint in PPS patients, occurring in over 60 percent of our series (Table 1). Two types of fatigue are reported by patients: generalized fatigue re-

| The University of Chicago: Post-Polio Clinic: Presenting Complaints in 97 Patients |
|---------------------------------|--------|--------|
| Symptom                        | Number | Percent |
| Pain                           | 75     | 77.3%  |
| Fatigue                        | 60     | 61.8%  |
| New Weakness                   | 54     | 55.6%  |
| New Atrophy                    | 15     | 15.4%  |
| Pain, Fatigue, Weakness        | 32     | 33.0%  |

Table 1.
quiring rest or sleep, and local muscle fatigue. Local muscle fatigue is most common in muscles previously severely affected by polio, and is often associated with cramps and fasciculations. Local fatigue may be a manifestation of ongoing muscle denervation, and is also reported by patients with classic denervating diseases such as amyotrophic lateral sclerosis.

Generalized (systemic) fatigue is common in PPS, but may also be a symptom of a variety of other states, including medical conditions such as diabetes mellitus, cardiopulmonary dysfunction, and thyroid disease. Depression ("low energy") may also lead to systemic fatigue. Once medical and psychiatric diseases have been ruled out, systemic fatigue in PPS may be a symptom of widespread neuromuscular junction transmission defects. We have found that patients with fatigue and marked increased jitter in single-fiber electromyography (an indicator of defective neuromuscular transmission) respond to agents which enhance neuromuscular transmission, such as the anticholinesterase pyridostigmine (Mestinon). Rest, ambulatory aids, and activity planning may also alleviate generalized fatigue.

New weakness is the third major component of the "post-polio triad" (Table 1). When new weakness occurs with new muscle atrophy, PPS patients are thought by some investigators to suffer from a specific syndrome of post-polio myelitis progressive muscular atrophy (PPMA).

It has been suggested that evidence of ongoing denervation (fibrillations and positive waves on EMG, increased jitter on single-fiber EMG, and atrophic muscle fibers on muscle biopsy) are diagnostic for this syndrome. However, we have found that electrophysiologic and muscle biopsy evidence of denervation is as common in polio patients who are not having new symptoms, as in patients who have clinically defined PPMA. Moreover, evidence of denervation is most severe in muscles which show the most signs of old polio. Thus, late denervation appears to be a concomitant of massive monophasic antecedent denervation, and not a sign of new disease. In addition, we found that although 14 out of 15 patients who complained of new atrophy also reported new weakness, only about one-half of patients who reported new weakness noted new atrophy. Thus, the relationship of atrophy to weakness is not clear.

New muscle weakness may put extra stress on a previously borderline compensated muscle, producing pain, cramping, and an "overwork myopathy," with accelerated weakness as an end result. It has been estimated that a partially denervated muscle graded "good" must work two and a half times as hard as normal muscle to accomplish the same task. We caution patients with new weakness to reduce activity. Exercise programs must be undertaken with extreme caution, and exercise should never be performed to the point of pain or muscle cramps. We advice patients to exercise limbs not previously affected by polio or, if this is impossible, participate in a carefully graded program in a therapeutically heated pool. One should exercise enough to prevent atrophy of disuse, but not enough to cause damage from overuse. High repetition, low resistance exercises are favored, as well as stretching and isometric drills. Orthotic devices, including the ankle-foot orthosis and knee-ankle-foot orthosis, may provide support for certain critically weakened muscle groups, although adequate function of other muscle groups (e.g., knee and hip extensor function for an ankle-foot orthosis) is a prerequisite for effective use. Wheelchair use should also be considered, sometimes only intermittently, as prolonged activity may predispose the patient to osteoporosis or venous thrombosis. Training in effective transfers, efficient movements, etc. by the physical and occupational therapist may also be useful, as can home help aids such as a shower chair and raised toilet seat.

Limb weakness may result in new joint instability, which in turn may be associated with new pain and increasing deformity. It has been noted, for example, that floor reaction with knee hyperextension serves a knee-locking function when the quadriceps is weak. However, profound degrees of weakness can provide a "positive feedback" situation where posterior knee ligaments are subjected to more torque stress, leading to further stretching. A knee-ankle-foot orthosis (fit with a posterior offset knee hinge) may prevent progressive joint damage in this situation.

Pulmonary complaints may occur in patients with previously weakened diaphragm, intercostals, abdominal, or accessory muscles. Frequently, a patient with previous paralytic poliomyelitis, involving muscles of respiration, will...
have borderline respiratory compensation for decades, and will undergo precipitous respiratory failure later in life. Increasing scoliosis, aspiration pneumonia, gradual loss of motor units with aging, and other factors may contribute to respiratory insufficiency. Respiratory symptoms (daytime somnolence, snoring, dyspnea, etc.) must be sought in all patients, particularly those with a history of respiratory involvement with polio. Baseline spirometry is also obtained in patients attending clinics. Muscle relaxants and medications which suppress respiratory drive should be avoided. Vaccines (pneumonia and flu vaccines) and cessation of smoking are also important in patient management. New respiratory muscle weakness may also present as sleep apnea, which may respond to medication (e.g., protriptyline), or may require night time oxygen or mechanical ventilation. Pulmonary complaints should always be evaluated and treated in conjunction with a pulmonary physician versed in neuromuscular diseases.

Prognosis of the post-polio syndrome depends upon the symptoms experienced by the patient and upon individual (as yet uncharacterized) differences in disease progression. General health care measures (proper rest, nutrition, weight management, etc.) as well as psychosocial support are important. Inflammation in joints and muscles may be managed well with the treatments cited above. At least some patients with PPS fatigue respond to anticholinesterase medications. Progressive weakness, with or without atrophy, is the least responsive symptom of PPS. Respiratory complaints, particularly, should be considered seriously. Fortunately, weakness progresses slowly (about one percent per year according to a recent study), and plateaus in function are observed. Although rapid progression of weakness does occur in some PPS patients, other diagnoses such as medical illnesses or superimposed neurologic and orthopedic problems must be considered.

A common complaint of post-polio patients is that health professionals do not understand or even believe their new symptoms. Although a breakthrough of understanding on PPS may not occur in the immediate future, it is the responsibility of all health personnel to listen carefully to patients with new problems and provide the best care possible. Specific symptomatic treatment should be made available where appropriate. The patient who has rehabilitated from the effects of acute polio must now be helped to accept the activity aids and lifestyle modifications necessary to ameliorate his "second disability."

ACKNOWLEDGMENTS

This paper was based on our experience at the University of Chicago Post-Polio Clinic, which was made possible by the fruitful collaboration of R. Maselli, R. Wollmann, R. Roos, E. Salazar, F. Brown, E. Nichols, R. Simon, P. Heidkamp, and R. Martia. We thank C. René de Cotret for preparing the manuscript.

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